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Bilateral Central Scotoma in a Middle-aged Man

Brittney Statler, MD; Sunil K. Srivastava, MD; Rishi P. Singh, MD

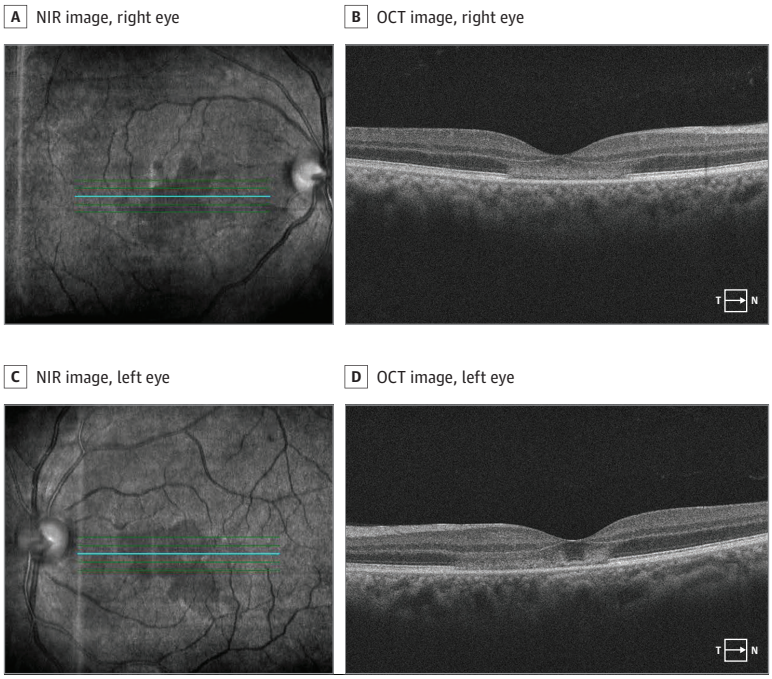

Figure. Near-infrared (NIR) imaging of the right eye (A) and the left eye (C) showed hyporeflectivity of the parafovea in both eyes. Optical coherence tomography (OCT) through a cross section of the affected area (horizontal blue line in A and C) of the right eye (B) and the left eye (D) revealed bilateral subfoveal disruption of the ellipsoid zone and outer nuclear layer. N indicates nasal; T, temporal.

A 43-year-old man presented as a referral for bilateral central vision loss. His medical history was significant for Crohn disease controlled with monthly injections of ustekinumab (Stelara; Janssen Biotech Inc). Symptoms of fever, headache, and myalgia began 1 week prior following a tick bite. He was prescribed oral doxycycline hyclate for suspected Lyme disease. Owing to persistent fever while receiving oral doxycycline hyclate, he was admitted for intravenous administration of doxycycline hyclate, and the fever resolved. He was found to be anemic (hemoglobin concentration, 6.7 g/dL; normal concentration, 13.5-17.5 g/dL) (to convert hemoglobin concentration to g/L, multiply by 10), requiring transfusion. While hospitalized, he developed acute bilateral central scotoma. Magnetic resonance imaging of the orbit and brain with and without contrast was unremarkable. Infectious serology test results for Lyme disease, syphilis, malaria, *Anaplasma* species, *Cryptococcus* species, Rocky Mountain spotted fever, and rickettsia were negative. Rheumatologic serology test results for antinuclear antibodies, including Sjögren anti-Ro and anti-La, anti-DNA, and anti-Smith, were also negative. Antineutrophil cytoplasmic antibody testing revealed elevated proteinase 3 antibody level. In the setting of anemia, infectious-disease consultants raised concern for babesiosis. Peripheral blood smear and polymerase chain reaction were performed; empirical therapy of azithromycin, 500 mg daily, plus atovaquone, 750 mg twice daily, was initiated. Polymerase chain reaction results for babesiosis later returned as negative.

Ocular examination revealed uncorrected distance acuity of 20/50 OD and 20/60 OS. Intraocular pressure values were normal, and there was no afferent pupillary defect. Slitlamp evaluation showed bilateral rare anterior vitreous cell. Dilated fundus examination revealed bilateral reddened foveal lesions and irregular macular pigmentation.

WHAT WOULD YOU DO NEXT?

- **A.** Obtain indocyanine green angiography
- **B.** Discontinue immunosuppression therapy
- **C.** Order optical coherence tomography
- **D.** Repeat neuroimaging

Diagnosis

Acute macular neuroretinopathy

What to Do Next

C. Order optical coherence tomography

Discussion

In this middle-aged man with bilateral central scotoma following recent illness, acute macular neuroretinopathy (AMN) was suspected. Near-infrared imaging showed hyporeflectivity of the parafovea, and spectral-domain optical coherence tomography (SD-OCT) revealed subfoveal disruption of the outer retina (**Figure**). Fundus autofluorescence and fluorescein angiography findings were unremarkable. A diagnosis of AMN was based on the examination and imaging findings. Findings of indocyanine green angiography (choice A) are typically normal in patients with AMN, and this procedure is indicated only if other causes of posterior uveitis are suspected. Immunomodulating agents (choice B) have not been implicated in AMN to our knowledge. Neuroimaging (choice C) is not indicated in AMN.

Acute macular neuroretinopathy was initially described in a 1975 case series[1] of 4 young women presenting with mildly reduced visual acuity, acute-onset paracentral scotomas, and red-brown, wedge-shaped macular lesions on ophthalmoscopy. All 4 women were taking oral contraception. Since then, environmental triggers reported include recent illness (47.5% of cases), oral contraception (35.6%), sympathomimetic substances such as caffeine and ephedrine (7.9%), septic shock (5%), and severe nonocular trauma (5.9%).[2] Acute macular neuroretinopathy exhibits sex, race, and age predilections, with young (mean age, 29 years) white women most frequently affected.[2] The process can be bilateral (55% of cases) or unilateral (45%), and scotoma is the chief concern in most cases (72.3%).[2]

Diagnosis of AMN is confirmed with ocular imaging, of which near-infrared and SD-OCT imaging are most sensitive.[3] Near-infrared imaging classically shows hyporeflective lesions at or near fixation, whereas SD-OCT reveals disruption of the ellipsoid zone (EZ) and hyperreflective disruption of the outer nuclear layer (Figure, D). In most cases (74%), fluorescein angiography findings are normal.[2] Underlying vascular abnormality is suspected in AMN,[4] and OCT angiography findings of flow voids within the deep capillary plexus and choriocapillaris have been reported.[5-7]

Acute macular neuroretinopathy should be distinguished from a more common OCT variant known as paracentral acute middle maculopathy, the lesions of which are more superficial than those of AMN, exhibiting hyperreflectivity within the inner nuclear layer and sparing of the EZ. Paracentral acute middle maculopathy is now understood to be an imaging finding in retinal vascular diseases such as diabetic retinopathy, retinal vascular occlusions, and retinal vasculitis; flow deficits within the superficial capillary plexus have been confirmed by OCT angiography.[4,8] Optimization of cardiovascular risk factors should be advised.

Individuals with AMN typically maintain visual acuity of 20/40 OU or better, with reduced yet persistent central scotoma. Optical coherence tomography hyperreflectivity fades over weeks to months, usually followed by thinning of the outer nuclear layer and/or EZ attenuation. Although treatment of AMN with corticosteroids has been reported,[9] most patients are observed.

Patient Outcome

In the setting of Crohn disease, we suspected an inflammatory cause for the bilateral central vision loss. This patient was prescribed 60 mg of oral prednisone daily; ustekinumab therapy was held. One month later, his uncorrected distance visual acuity improved to 20/25 OU, with subjective improvement of central scotomas. Disruption of the outer nuclear layer on OCT diminished, leaving behind residual EZ attenuation. A slow tapering of the oral prednisone dose was initiated.

ARTICLE INFORMATION

Author Affiliations: Cole Eye Institute, Cleveland Clinic, Cleveland, Ohio (Statler, Srivastava); Center for Ophthalmic Bioinformatics, Cole Eye Institute, Cleveland Clinic, Cleveland, Ohio (Singh).

Corresponding Author: Rishi P. Singh, MD, Cole Eye Institute, Cleveland Clinic, 9500 Euclid Ave, I-32, Cleveland, OH 44195 (singhr@ccf.org).

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